FAMILIAL SYNKINESIS IN OCULO-PALPEBRAL MOVEMENTS*

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SEVERAL isolated examples of congenital ptosis showing synkinetic oculo-palpebral phenomena are recorded in the literature. After an unsuccessful search for examples of hereditary synkinesis, the author reports the cases of two sisters with congenital ptosis who showed abnormal ocular movements.

Case Reports

Case 1, a female aged 38 years, had a partial congenital ptosis on the right side and an unduly wide palpebral fissure, amounting to a lid retraction, on the left (Fig. 1). Elevation of the right eye was completely absent and that of the left was markedly restricted (Fig. 2). On looking down, only the left eye moved downwards in a normal manner, while the right eye diverged (Fig. 3). The left upper eyelid followed the ipsilateral ocular movement in a normal manner but the right upper eyelid did not move downwards and the palpebral aperture remained open, simulating a pseudo-Graefe phenomenon of lid retraction on downward movement of the eyes (Fig. 4). Lateral versions and convergence were normal (Figs 5 and 6) and a general neurological examination revealed no other abnormality.

FIG. 1.—Ptosis on right side. Lid retraction on left side with a wider palpebral aperture in the primary position.

FIG. 2.—Absence of elevation.

FIG. 3.—Abduction of right eye on looking down.

FIG. 4.—Pseudo-Graefe phenomenon on right side on looking down.

FIG. 5.—Normal dextro-version.

FIG. 6.—Normal laevo-version.

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Case 2, a female aged 33 years, the younger sister of Case 1, had a congenital bilateral ptosis (Fig. 7). There was no lid elevation on contraction of the frontalis muscle. Both eyes were hypotropic to the extent of 15° and elevation was completely absent. On attempting elevation there was a tendency to convergence (Fig. 8). Downward movement was restricted and on looking down the left eye diverged (Fig. 9). Lateral versions and convergence were normal and a general neurological examination showed no further abnormality.

FIG. 7.—Bilateral ptosis in primary position.

FIG. 8.—Convergence on elevation.

FIG. 9.—Abduction of left eye on looking down.

Summary

Two sisters with congenital ptosis are reported. A familial influence in the aetiology of the synkinetic oculo-palpebral phenomena, rarely reported, is noteworthy. Several features were common to both cases. Versions were normal, elevation was absent or grossly restricted, and looking down caused abduction of one eye in both cases. In Case 1, downward movement of the eyes led to an appearance simulating the pseudo-Graefe phenomenon. Case 2 showed convergence on attempted elevation.